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# Pacemaker Treatment of Sick Sinus Syndrome in Children

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**The sick sinus syndrome is being recognized with increasing frequency in children. Although it is sometimes benign, it can be serious or have fatal consequences. Fifty-one patients (mean age 10.5 years) underwent permanent cardiac pacing for sick sinus syndrome. Twenty patients had epicardial ventricular pacing and 12 had an epicardial atrial implant. Seven had endocardial atrial pacing, six epicardial atrioventricular (AV) sequential pacing, four epicardial universal pacing and two endocardial universal pacing.**

**Of the 49 symptomatic patients, 45 had relief of symptoms. Eleven of 18 patients with associated tachyarrhythmias had amelioration of their tachycardia. There were no early but two late deaths unrelated to the pacemakers. Seven patients during a mean follow-up period of 26 months required reoperation for pacing lead or sensing problems. Permanent pacing for sick sinus syndrome in children is a safe and symptomatically effective procedure.**

The sick sinus syndrome is being recognized with increasing frequency in patients in the pediatric age range (1–7). The syndrome is defined as electrocardiographic or electrophysiologic evidence of sinus node dysfunction, including sinus bradycardia, sinus arrest and sinus exit block. Tachyarrhythmias are often present but are not required for the diagnosis. This is true not only in patients who have undergone palliative or reparative operation for congenital heart defects, but also in patients without surgical treatment for congenital heart defects or cardiomyopathies, those being treated with antiarrhythmic drugs and patients with an otherwise normal heart.

Although some patients with sick sinus syndrome are asymptomatic, many symptomatic patients as well as a few who have died (8–10) have been described. In this paper, we report the techniques and results of permanent cardiac pacing in pediatric patients with the sick sinus syndrome. This group includes each patient paced for sick sinus syndrome from 1976 to 1981 at Texas Children's Hospital. The indications and techniques have evolved over this period.

## Methods

**Patients.** From January 1, 1976 to December 31, 1981, we prospectively evaluated all pediatric patients with symptoms of syncope, fatigue or congestive heart failure for evidence of sinus node dysfunction. We used 24 hour ambulatory monitoring (11), treadmill exercise electrocardiography (12) and invasive determination of sinus node function (13). We reviewed the records of 51 patients in this study who had received an implanted cardiac pacemaker and paid particular attention to age, anatomic diagnosis, electrocardiographic diagnosis and electrophysiologic variables. We then reviewed the types of pacemakers and leads used, as well as the surgical approach and results of cardiac pacing in this group of patients.

During this period, we performed intracardiac electrophysiologic evaluation on 96 pediatric patients with known or suspected sick sinus syndrome. We used standard electrophysiologic techniques previously reported from our laboratory (3,13,14).

**Indications for cardiac pacing.** Pacemakers were implanted primarily for symptoms of syncope or near syncope. They were also implanted for treatment of congestive heart failure or excessive fatigue, as well as for help in controlling

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supraventricular and ventricular arrhythmias. The charts of these patients were reviewed to determine the results of cardiac pacing.

## Results

**Patient characteristics.** Fifty-one patients ranging in age from 2 to 29 years (mean 10.5) had permanent pacing for sick sinus syndrome. Thirty had had surgery for congenital heart defects (Table 1). In five patients, the sick sinus syndrome was induced by drugs used in treating tachyarrhythmias (Table 2). Four patients had congenital cardiac defects but had not undergone surgery. Two patients had congestive cardiomyopathy and 10 had no associated heart disease. Eighteen had associated tachyarrhythmias (14). Forty-nine underwent pacing because of symptoms and two because of heart rates that were considered unacceptably slow.

*Forty-eight patients had electrophysiologic studies before pacemaker implantation.* Thirty-five had an abnormally long corrected sinus node recovery time and nine had a prolonged sinoatrial conduction time. Atrioventricular (AV) conduction was entirely normal in 30, mildly abnormal in 9 and severely abnormal in 12.

**Mode of pacing.** The mode of pacing was recommended on the basis of electrophysiologic findings, but practical considerations were also taken into account. It was considered optimal to use atrial demand (AAI) pacing for sick sinus syndrome with normal or near normal AV conduction. However, during the first several years of this study, a thoracotomy was required to attach an atrial lead while a ventricular lead was attached by a subxiphoid or subcostal approach.

*Twenty patients had an epicardial ventricular demand pacemaker while 12 had atrial epicardial implantation.* In each patient with an epicardial implant, the pulse generator was implanted beneath the rectus muscle. Each ventricular implant was by a subxiphoid or subcostal approach. Nine atrial implants were by thoracotomy (seven left and three right) and three were by a new subxiphoid technique (Fig. 1). Seven patients received endocardial atrial pacing (Fig. 2 and 3), six epicardial AV sequential, four epicardial universal and two endocardial universal pacing (Fig. 4). Each patient who had endocardial leads placed had the pulse generator implanted subcutaneously in the prepectoral fascia. Atrial, ventricular and AV sequential pulse generators were standard programmable units from Cardiac Pacemakers Incorporated, Intermedics and Medtronic. The six universal AV squential pulse generators were investigational Medtronic units.

*Ventricular epicardial leads* were Intermedics or Medtronic two-turn sutureless models. Atrial epicardial leads were Cordis nine-turn (9) or Medtronic stab-on sutureless (3). Ventricular endocardial leads were Intermedics or Medtronic tined polyurethane. Atrial endocardial leads were Intermedics or Medtronic tined polyurethane "J" or Medtronic screw-in models.

**Operative results.** There was no mortality associated with pacemaker implantation. Two patients died after hospital discharge, one patient at 1 month and one at 3 months. Each pacemaker was working at the time of death. Both patients had severe residual congenital heart disease and depressed cardiac muscle function.

**Follow-up results.** The follow-up period ranged from 6 months to 5.5 years (mean 2.4 years). Forty-five patients had relief of their symptoms and four did not. Three of the patients with residual symptoms had tachyarrhythmias. One had pacemaker syndrome (that is, lightheadedness at the onset of ventricular demand pacing).

Eleven of the 18 patients with tachycardias had improvement in their tachycardia control concomitant with pacemaker implantation. Tachycardia was unchanged in five patients and worse in two. In these two, accessory connections were present and were successfully divided in each.

*Three patients with an epicardial atrial implant had to undergo reoperation* after 3, 4 and 5 years, respectively, because patient growth caused electrode displacement. One patient with an atrial endocardial screw-in lead had to have reoperation 1 month after initial implantation because of

**Table 1.** Postoperative Congenital Heart Disease in 30 Patients

| Patients (no.) | Lesion | Operation | Age at Operation (yr) | Age at Symptoms (yr) |
|---|---|---|---|---|
| 15 | d-TGA | Mustard | 4.5 | 10.1 |
| 7 | ASD-2 | Closure with CPB | 7.6 | 12.3 |
| 3 | ASD-1 | Closure with CPB | 8.0 | 8.6 |
| 2 | Common atrium | Separation with CPB | 9.7 | 9.7 |
| 3 | Other | Repair with CPB | 15.5 | 16.0 |

ASD-1 = primum type atrial septal defect; ASD-2 = secundum type atrial septal defect; CPB = cardiopulmonary bypass, d-TGA = d-transposition of the great arteries

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**Table 2.** Drug-induced Sick Sinus Syndrome in Five Patients

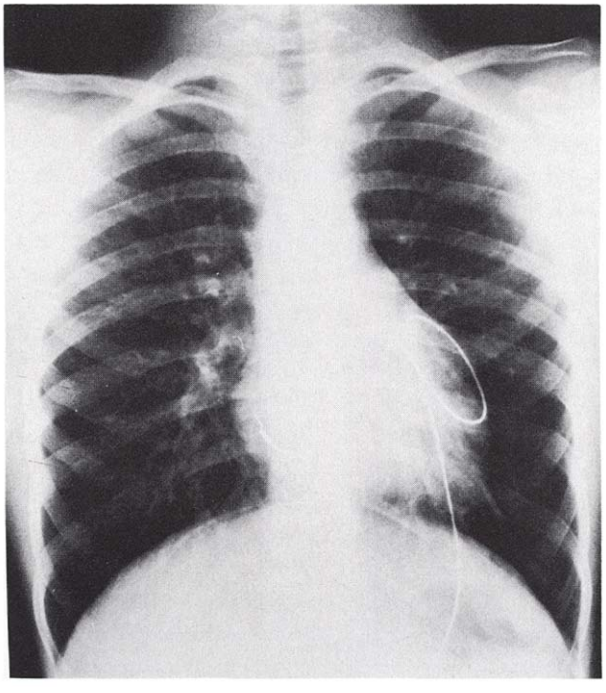
| Case | Tachyarrhythmia | Drugs | Result |
|---|---|---|---|
| 1 | SVT, right Kent bundle | Dig and Q, P and Q | Worse, Kent bundle division |
| 2 | AVN reentrant SVT | Dig and P, dig and V | Asymptomatic |
| 3 | SVT, left Kent bundle | Dig and Q, P and Q | Worse, Kent bundle division |
| 4 | AVN reentrant SVT | Dig and V | Asymptomatic |
| 5 | AVN reentrant SVT | Dig and V, Q, dig and Q, Q and P | Improved |

AVN = atrioventricular node; Dig = digoxin, P = propranolol, Q = quinidine, SVT = supraventricular tachycardia, V = verapamil.

phrenic nerve stimulation. Three patients with a ventricular epicardial lead and a ventricular demand pulse generator had to have reoperation, two because of sensing problems and one because of pacemaker syndrome. The two sensing problems were corrected by implanting units with programmable sensitivity using the same leads. The pacemaker syndrome was treated with an AV sequential unit using the same lead. Thus, none of the ventricular epicardial leads malfunctioned.

*Follow-up time on endocardial leads* ranged from 6 to 18 months, whereas follow-up on epicardial leads ranged from 9 months to 5.5 years. Each pulse generator is currently set at 5 V and 0.6 ms or less; none of the leads has developed a "high threshold."

**Figure 1.** Posteroanterior chest radiograph of a patient who had Mustard's operation for d-transposition of the great arteries. Two years postoperatively, she had severe sinus bradycardia and underwent implantation of an atrial demand pacemaker. A left thoracotomy was used to attach the plaque type sew-on lead to the base of the left atrial appendage. Four years later, the lead failed to capture the atrium and a "stab-on" atrial lead was placed by a subxiphoid approach.

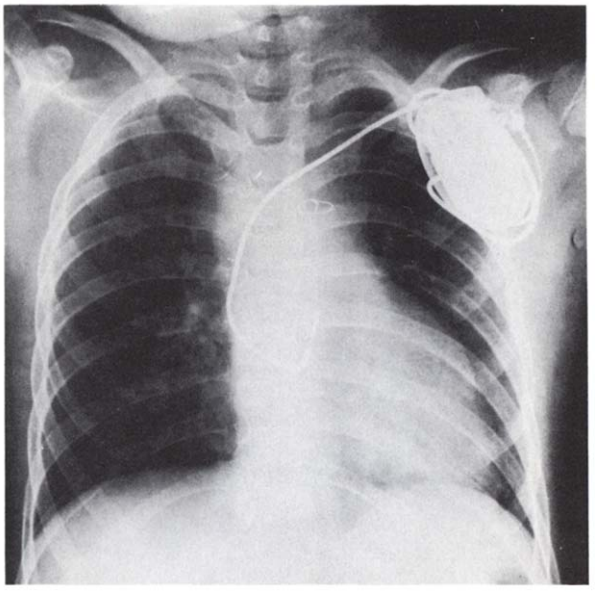


## Discussion

Although the sick sinus syndrome occurs most commonly in the elderly (15), it is being recognized with increasing frequency in the pediatric age group (1–7). This is true not only because the most common cause is cardiac surgery, but also because there is an improved degree of electrophysiologic understanding among pediatric cardiologists.

The sick sinus syndrome may be asymptomatic in some patients, while in others it may cause symptoms or even death (8–10). The histopathologic features of the sick sinus syndrome have been studied (8–10). Destruction and fibrous replacement of the sinus node as well as fibrosis of the

**Figure 2.** Chest radiograph showing an atrial demand pacemaker with a polyurethane tined "J" lead placed by a left subclavian vein puncture. This 5 year old girl had Mustard's operation for transposition of the great arteries.



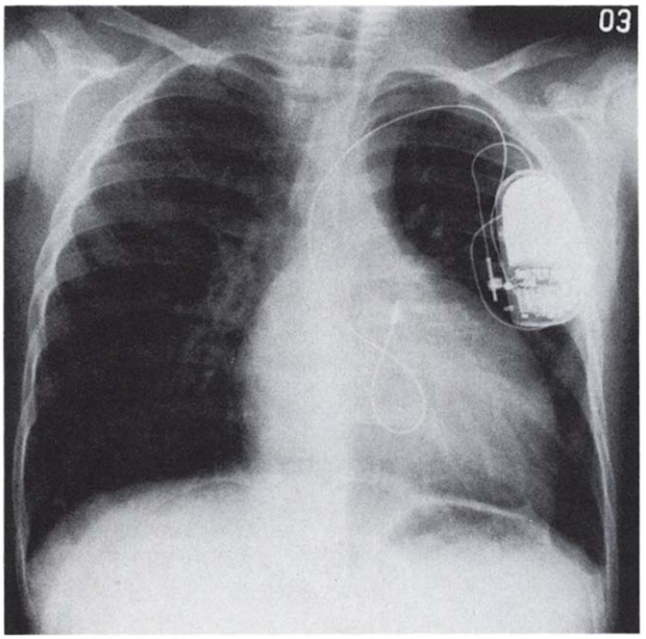



**Figure 3.** Chest radiograph showing the use of an endocardial screw-in lead with an atrial demand pacemaker in a 9 year old child after Mustard's operation. The lead is screwed into the roof of the anatomic left atrium and a loop is left in the atrium to try to accommodate growth.

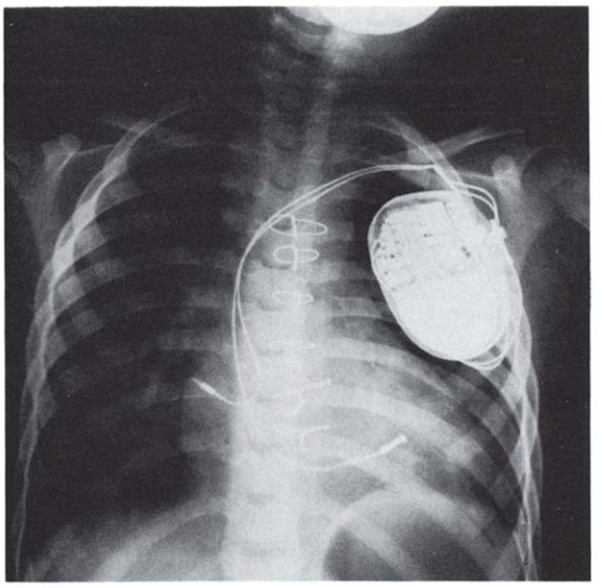


**Figure 4.** Universal atrioventricular (AV) sequential pacemaker implanted by transvenous technique in a 4 year old child after repair of complete AV canal defect. A screw-in lead is used in the atrium and a tined lead is used in the ventricle. The child weighed 16 kg. The crimp in the leads is where they are tied near their insertion in the left subclavian vein.

approaches to the sinus node have been found (8). Abnormalities of the atria and approaches to the AV node were also observed.

**Indications and advantages of atrial pacing.** In adults, the treatment of symptomatic sick sinus syndrome is cardiac pacing (15). Ventricular demand pacing was the initial mode used. As technology of pacing has improved, atrial, AV sequential and AV universal pacing have been used (16,17). Because many patients with sick sinus syndrome have intact AV conduction, atrial pacing would seem to be the most appropriate treatment. Controversy exists, however, because of concern as to possible later onset of AV conduction defects. In our patient group, we have not seen the development of AV conduction defects in any patient with an atrial pacemaker. These patients have been followed up for as long as 5.5 years. In children with surgical sick sinus syndrome, the degree of AV conduction abnormality is probably fixed by the damage during surgery. Although many of our atrial implants were in patients with surgical sick sinus syndrome, we also used atrial pacemakers in patients with other causes of sick sinus syndrome including cardiomyopathy. Our longest duration of an atrial implant in a patient with cardiomyopathy is 4 years. We used electrophysiologic studies to document any abnormalities of AV conduction before pacemaker implantation. Atrial pacing offers an advantage over other pacing modes because the two ventricles contract nearly simultaneously if there is no bundle branch block. In addition, it requires use of only one lead, and a small, relatively simple pulse generator may be used. Thus, we believe that there will be a continuing place for atrial demand pacing in patients with sick sinus syndrome.

**AV sequential pacing.** Patients with sick sinus syndrome and associated AV block are candidates for some mode of AV sequential pacing. Standard AV sequential pacemakers do not sense atrial activity; thus, patients cannot increase their ventricular rate even if their sinus node can respond to exercise. Most pediatric patients with sick sinus syndrome can increase their heart rate with exercise (12). In addition, competitive atrial rhythms may be induced by AV sequential pacing because of lack of atrial sensing.

*In light of these problems, we evaluated a fully automatic AV sequential pacemaker.* This pulse generator has allowed the most physiologic response in patients with combined sick sinus syndrome and AV block. However, this mode of pacing has also given rise to a new type of complication: pacemaker reciprocating tachycardia. This only occurs in patients with intact retrograde conduction. When a premature ventricular complex conducts retrograde to the atrium, the pacemaker may sense the atrial depolarization and stimulate the ventricle, thus establishing a reentrant circuit. This does not occur in every patient with retrograde conduction. The only way to treat this tachycardia with currently available pacemakers is to reprogram to AV sequential mode.

Pacemaker reciprocating tachycardia did not occur in any patient in this series.

*Pacing in sick sinus syndrome involves an atrial lead in most patients.* Our techniques of atrial lead implantation have evolved from direct implantation by thoracotomy to a subxiphoid approach and finally to transvenous implantation. The size of pulse generators, even AV universal, is now acceptable for transvenous implantation in children weighing more than 15 kg. Active fixation leads make it possible to use transvenous implantation even in patients who have undergone previous open heart surgery, including the Mustard operation.

Appropriately selected pacing systems allow the patient with sick sinus syndrome to lead a normal or near normal life in the short term. Multiple reoperations will be necessary.

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